



Huntington's Disease Society of America

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Presenter Disclosures

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The following personal financial relationships with commercial interests relevant to this presentation existed during the past 12 months:

Consultant to Lundbeck
Speaker for Allergan



HD411:

An Introduction to Huntington's Disease

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Goals for Today

- Review what problems people with HD have
- Talk about some facts and myths
- Answer your questions

George Huntington (1850-1916)

- Family physician
- Worked on Long Island
- Originally read his report to a group of family physicians in Ohio on February 15, 1872



How Many People Have HD in the US?

- 3,000
- 30,000
- 300,000
- 3 million
- 3 bazillion (almost a google)

What is a Gene?

- A comfortable pair of pants
- The guy who lives on your street
- Billy ____ from Michael Jackson's song
- The tiny part of our body that is the blueprint for making the building blocks for the rest of our body
- A treatment for Huntington's disease

Who Gets HD?

- Boys and girls both affected
- All races and religions
- HD usually starts when people are in their 30's or 40's
- Some people show signs of the disease early
 - About 1 in 10-20
 - Called juvenile HD if it starts before age 20

Genetic Disease

- Each child of an affected parent has 50/50 chance of inheriting disease (like the flip of a coin)
- Does not skip generations
- If you do not inherit the gene, you cannot pass it to the next generation

Three Main Problems with HD

- Trouble with:
 - Moving
 - Thinking
 - Behaving

Movement Disorder

- Uncontrolled movements
 - Dance-like movements (chorea)
 - Twisting or extra muscle contractions (dystonia)
 - Slow movements and stiffness (parkinsonism)
- Slurred or irregular speech
- Trouble swallowing
- Coordination problems
- Walking and balance problems

Thinking Problems

- Slow thinking
- May make bad decisions
- Poor attention

Behavior

- About half of patients with HD may have sadness or nervousness
- Thinking can get stuck on 1 thing
- Can believe things that are not true
- Personality changes
- Bad sleep
- Trouble controlling themselves

How Do We Measure Disease Progression?

- Total Functional Capacity (TFC) scale
 - Work
 - Handling money
 - Doing things around the house
 - Caring for themselves
 - Where are they living?
- Not very good for kids, people with behavior problems or later stages of disease

Myth or Fact?

- Huntington's disease is diagnosed with a blood test for the gene
 - Myth
 - Fact
 - I have no idea

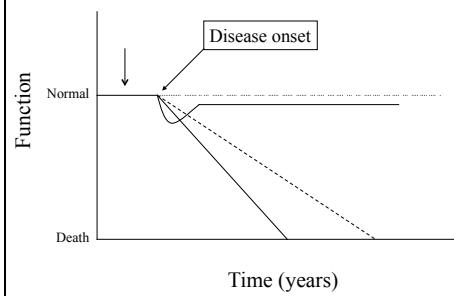
Diagnosis

- Based on story that patients tell me about the problems they have been having, their family problems and what I see on their exam.
- Sometimes I order other tests like:
 - MRI, CAT scan, EEG or
 - Gene test

When to Think About Getting the Gene Test

- "At risk" adult patient without symptoms
 - Family planning
 - Financial planning
 - Need to know
- When someone has the disease, but no one else in the family does
 - Adopted
- Juvenile case (when showing problems likely to be the disease)
- Unusual symptoms

Changing Disease Progression

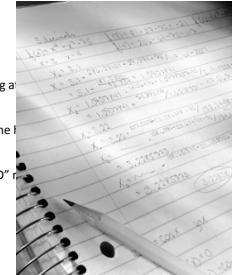


Treatments

- Movements
- Depression or nervousness

Hints to Help Patients with Thinking Problems

- Memory
 - Picking from a list is easier
 - Give hints, yes/no questions
- Learning
 - Give simple, clear instructions
 - Limit distractions - focus on 1 thing at a time
- Organization
 - Make lists
 - Keep a big, common calendar in the living room
 - Routines
- Slow thinking
 - No response does not mean a "NO" response
 - Have patience



Treatments – Not Just Medicines

- Social work
- Nutrition
- Social support
- Occupational therapy
- Physical therapy
- Speech therapy

Questions

- I am confused about something and have a question.
 - (that means everybody else probably is too – just ask)
- I do not have any questions.
- I have a question but am too shy to ask.
- I am overwhelmed and need to think about this for a little while.